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DOI: https://doi.org/10.1017/s0022215100046533

The Journal of
Laryngology and Otology

(*Founded in* 1887 *by* MORELL MACKENZIE *and* NORRIS WOLFENDEN)

June 1949

HISTOPATHOLOGICAL INVESTIGATIONS ON THE LOCALIZATION, NUMBER, ACTIVITY AND EXTENT OF OTOSCLEROTIC FOCI*

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IT is considered that otosclerotic foci are localized to certain sites of predilection, i.e. to the window regions and particularly to the area anterior to the oval window, in the latter case often accompanied by ankylosis of the stapes. Although the sites of predilection of the foci have long been known, only few papers are to be found in the literature regarding their distribution in the whole bony capsule of the labyrinth in large series (Nager, Nager and Meyer, Guild).

In the histological examination of material with clinically established otosclerosis most authors have found that nearly 100 per cent. of the foci are located in the oval window region and are accompanied by ankylosis (Manasse, Mayer, Wittmaack, Lange, Kosokabe, Weber, Nager, Nager and Meyer, and others). In *frequency* investigations of otosclerosis in unselected material, this figure decreases considerably, and stapes ankylosis is less often encountered (see especially Engström and Guild). Relatively little appears to be known about the localization of the lesions in the labyrinthine capsule outside the region of the stapes. This is partly due to the fact that in many cases foci in other regions than those of the windows have not been sought.

It, therefore, appeared to the present writer to be of value to investigate the localization of foci in the whole bony capsule of the labyrinth

* This paper will be reported in more detail in *Upsala Läkareför. Ny följd.*, Vol. 54, 1-2, 1949.

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on a material of temporal bones with otosclerosis. In addition, a study of the number of foci, their activity and extent seemed to be of considerable interest. More complete knowledge of this subject than we possess at present could possibly lead to the solution of the problem of the ætiology of otosclerosis or at least provide suggestions for further investigations.

In a later paper the clinical aspects of the material will be dealt with and the relation discussed between the clinical and the histological findings and—to some extent—of therapeutical measures on the basis of the results obtained.

The *material* in the present investigation consists of the collection of temporal bones assembled by Nager over a period of many years. The cases are partly clinical otosclerosis cases, partly cases of suspected otosclerosis and partly cases where there were no suspicions of the disease.

The material examined consisted altogether of seventy-four cases with typical otosclerosis (121 otosclerotic bones) and an additional three cases with pathological bone changes ("Umbau ") only. One of these seventy-four cases was found in a closer study of fifty unselected cases of acute and chronic otitis media, with or without cholesteatoma or other complications, among which I also found a case of " Umbau " in the cochlear capsule. In addition I investigated eighteen cases of tuberculosis of the ear and found unilateral pathological bone changes in one. In fifteen cases of tumour of the acusticus there were similar bilateral changes in the stapedial footplate in one instance. No otosclerotic changes or " Umbau " were found in the following : eight cases of syphilis, thirty of diseases of the bones (Paget's disease, von Recklinghausen's disease, osteogenesis imperfecta, etc.). In twenty-five cases of deaf-mutism there were five cases of otosclerosis.

The histological treatment of the specimens was uniform throughout, i.e. fixation, decalcification etc. The thickness of the sections was usually about 20μ and approximate calculations of the extent of the foci at right angles to the direction of the section were made on this figure.

To make clear the location of each focus in the labyrinthine capsule, this is indicated on a model of the labyrinth taken from one of *Siebenmann's* works (v. Fig. 2). The degree of activity of the foci is divided into active, quiescent and mixed, the characteristics of the different stages being those usually given (v. Nager).

In every case data concerning the location, activity, extent and number of foci as well as all available clinical and histological data were recorded and assembled in tabular form. Thirty-six of the cases have been reported earlier by Nager, Nager and Meyer, and others ; the other thirty-eight being described here for the first time.

Histopathological Investigations on Otosclerotic Foci

As the *result* of this investigation the following statement can be made: a careful distinction must be made between the clinical and the histopathological diagnoses, a fact pointed out by many earlier writers.

Of the seventy-four cases investigated thirty-four were females and forty males; the quotient of the ratio females: males =0·85:1. The present material is too small to permit any conclusions regarding the incidence of otosclerosis in men and women. Moreover, only a small proportion of the material consists of a random selection of temporal bones (et. Engström and Guild). The findings are nevertheless interesting

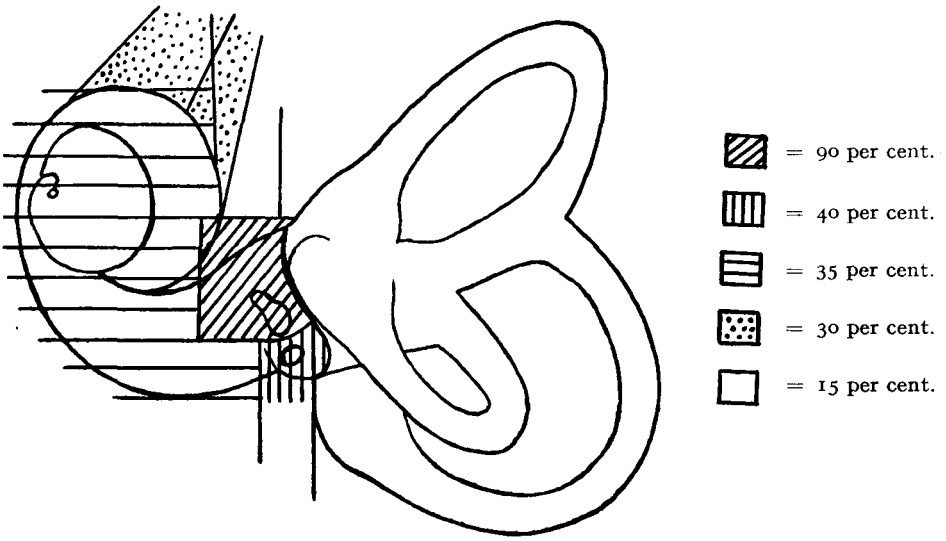

FIG. 1.
LOCALIZATION OF THE OTOSCLEROTIC PROCESS IN THE LABYRINTHINE BONE CAPSULE (schematically).

and indicate to some extent that so-called histological otosclerosis is as common in men as in women. This is supported by the results of earlier histological investigations by Lange and Engström.

The localization of the otosclerotic process in the bony capsule of the labyrinth is seen schematically from a model of the labyrinth (v. Fig. 1). In 90 per cent. of all cases the localization is in the oval window region, 50 per cent. being accompanied by stapes ankylosis. In 40 per cent. the process is localized to the round window region. The cochlear capsule is involved in 35 per cent., the internal auditory canal region in 30 per cent. and the semi-circular canal capsule in 15 per cent. The investigation thus shows that in addition to the typical localization of the otosclerotic process to the window regions, other parts of the labyrinthine bony capsule are involved in a large number of cases.

Typical otosclerotic foci entirely outside the bony capsule were found in two instances only. In the literature only few reports have described otosclerotic lesions outside the actual capsule (Cowell, Nager, Kelemen, Ruttin) and they are considered to be very rare. In both cases reported here the otosclerotic process was located to the ossicles (these cases earlier reported by Cowell and Nager).

The otosclerotic process was limited to only one of the regions of the capsule in 55 per cent. of the whole material. It was then in the stapedial region in 40 per cent. of all cases, and ankylosis was present in 18 per cent. In two instances the focus was confined to the stapedial footplate. In 8 per cent. of the entire material the process was confined to the round window and a single focus at another site in the capsule than the aforementioned was rare (altogether 7 per cent.).

The distribution according to sex is usually in direct relation to the number of males and females in the material. An exception is found if the number of cases with foci at the oval window accompanied by stapes ankylosis is compared with those without the latter. Contrary to expectation there was a much higher proportional incidence of ankylosis among females. Since stapes ankylosis gives rise to clinical symptoms, the disease should then be manifest clinically in a higher frequency in women, whereas, as mentioned previously, the disease on histological examination is as frequent in men. That the disease clinically is found more often in women, could be explained by the fact that stapes ankylosis occurs more often in the case of the former. Nevertheless, as already pointed out, no definite conclusions can be drawn in this respect owing to the relatively small size of the material, but it is of interest to note the tendency.

No difference was apparent in the entire material as regards the incidence in the right and the left bones. The pathological areas were usually symmetrically located in both temporal bones, unilateral otosclerosis being an obvious exception. The material consisted of cases from 16 years of age to 87. Owing to the relatively small number of cases, the material was divided into two groups only, one over fifty years and the other fifty years or less. No difference as regards the localization appeared between them.

As did many other writers I found a single focus in approximately 65 per cent. of the total number of bones ; in the others there were two or more foci. More than three foci were rare. The number of foci appeared to decrease with increasing age.

As regards the activity, I found that active foci occurred in 20 per cent. of the cases, quiescent in 30 per cent. and mixed in approximately 50 per cent. When several foci were present in the same bone or in the same individual, they were usually of the same degree of activity. In the present material there were no instances of active foci only over the age of 50 year, when mixed and quiescent areas were more common. The writer

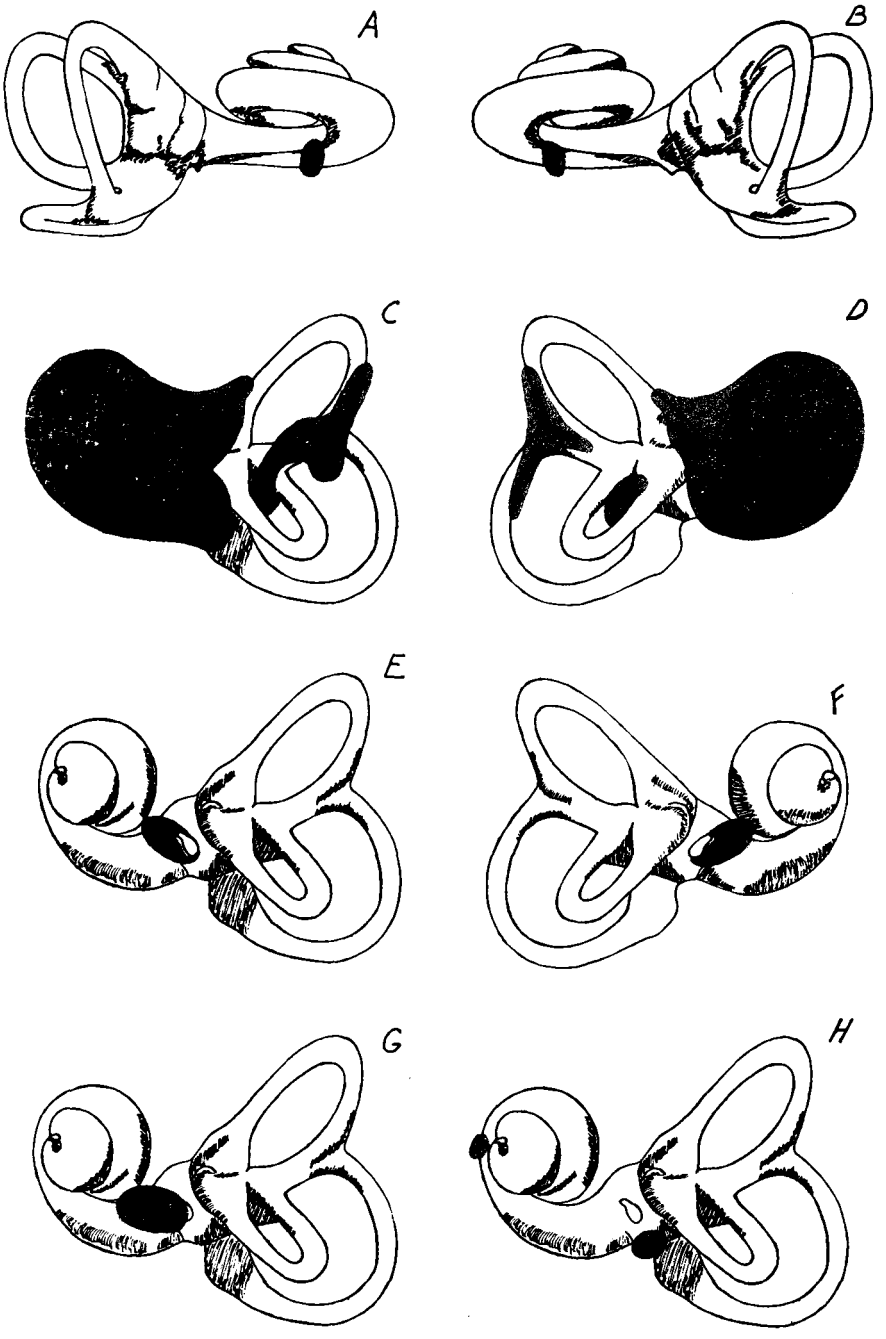

Fig. 2.

Some Cases of Otosclerotic Foci with a Typical Extent in the Labyrinthine Bone Capsule.

2A and B are Case 15, left and right temporal bones, C and D Case 43, left and right temporal bones, E Case 44, left temporal bone, F and G Case 49, both temporal bones, and H Case 54, left temporal bone.

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suggests that this fact can be of clinical interest, since it indicates that such cases with a more quiescent or arrested process can be suitable subjects for operation (fenestration) in view of more lasting results.

The extent of the process varied considerably, from 0·4 mm. to more than 20 mm. There were no demonstrable correlation between the extent, the localization and the activity.

In the material I found so-called diffuse otosclerosis in approximately 10 per cent. of the cases. The process was then usually very extensive, the entire cochlear capsule, the vestibule and even parts of the semicircular canal capsule generally involved. Stapes ankylosis was considerably more common in comparison with the material as a whole and the activity was extremely mixed.

In earlier histological investigations, unilateral otosclerosis was stated to occur only exceptionally, and it is in recent works that this form of the disease has been shown to be not uncommon. Nager and Meyer found an incidence of approximately 15 per cent., but Guild in his series of unselected material gives a figure as high as approximately 30 per cent.

In the present material a definitely unilateral localization of the otosclerotic process was found in sixteen cases out of sixty-three, i.e. in somewhat more than 25 per cent. This finding in the present investigation as well as in other recent investigations indicates that unilateral otosclerosis occurs in 15-30 per cent. of the cases in collections of histological specimens. The extent of the foci is often small and they are usually localized to the window regions. Foci at the oval window without stapes ankylosis are very common in these cases.

Histologically, the foci in the oval window region were typical in extent. The round window was often occluded. The lumina of the cochlea, the vestibule, and the semicircular canals were often compressed, particularly in cases of diffuse otosclerosis, whereas foci in the internal auditory canal did not appear to compress the lumen to any extent. There were six instances of otosclerotic bone formation in the basal coil of the scala tympani. Smaller foci (< 3 mm.) are usually located in the periosteal bone layer. It has been considered that the enchondral bone layer is the site of the new formation of bone ; the results of the present investigation nevertheless distinctly contradict this theory.

Inflammations of the ear, such as otitis media with or without complications were found in approximately 10 per cent. of the cases. Other types of bone disease concomitant with otosclerosis were rare with the exception of endemic deaf-mutism. Five cases of otosclerosis showed changes more or less typical of this disease. Atrophy of the neural elements is uncommon. If it is present there is usually a tumour of the acusticus.

Pathological bone changes (" Umbau ") were observed in many cases. They were found to be fairly common in the semicircular canal capsule

(eleven cases of seventy-four) and more uncommon in the cochlear capsule and the vestibule (four cases of seventy-four). These changes were also found in the ossicles. The theory that the pathological bone changes are a preliminary stage of otosclerosis has been put forward (Weber, and others). This hypothesis is not supported by the present writer's findings, since typical otosclerotic lesions in the semicircular canal capsule occurred in 15 per cent. only, whereas the cochlea and the vestibule were involved in 35 per cent., i.e. a diametrically opposed condition to the occurrence of pathological bone changes (" Umbau "). One explanation could be that the typical otosclerotic lesions have no connection with these pathological bone changes but they are entirely independent processes in the same bone. In order to solve this problem, more thorough investigations are required, particularly with regard to the nature of the so-called Umbau.

Summary

A report is given of an investigation on the localization, number, activity and extent of otosclerotic foci in seventy-four cases examined histologically (121 otosclerotic bones). It is pointed out that the otosclerotic process in addition to the typical localization to the window regions, also involved other parts of the labyrinthine bony capsule in a large number of instances (v. Fig. 1, page 323). The fact that in the investigated material as well as in other reports, otosclerosis in men seems as frequent as in women is discussed and it is pointed out that, at least in this material, stapes ankylosis is more common among women than among men. Active areas in cases over fifty years of age appear to be uncommon. In histological series unilateral otosclerosis is usually found in 15-30 per cent. of the cases. Finally, the nature of pathological bone changes (" Umbau ") is discussed.

The writer wishes to express his sincere gratitude to Professor F. R. Nager for the valuable material which he placed at the author's disposal, and for the great interest with which he has followed the progress of this work.

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